

WHAT IS CLAIMED IS:

1. A method of enhancing the activity of lysosomal α -galactosidase A in mammalian cells comprising administering an effective amount of a compound selected from the group consisting of 2,5-dideoxy-2,5-imino-D-mannitol, 3,4-diepi- α -homonojirimycin, 5-O- α -D-galactopyranosyl- α -homonojirimycin, 1-deoxygalactonojirimycin, 4-*epi*-fagomine, calystegine A₃, calystegine B₂, and calystegine B₃, and N-alkyl derivatives thereof.

514/317 (US Pat 5,536,732)

2. The method of claim 1 wherein the lysosomal α -galactosidase A is a mutant form which is present in patients with Fabry disease.

3. The method of claim 1 wherein said cells are human cells.

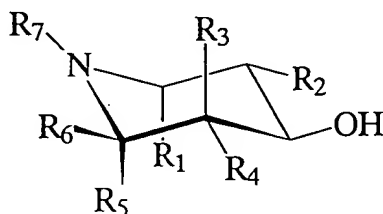
4. The method of claim 3 wherein said cells are the cells of a patient with Fabry disease.

5. A method of treating Fabry disease comprising administering an effective amount of a compound selected from the group consisting of 2,5-dideoxy-2,5-imino-D-mannitol, 3,4-diepi- α -homonojirimycin, 5-O- α -D-galactopyranosyl- α -homonojirimycin, 1-deoxygalactonojirimycin, 4-*epi*-fagomine, calystegine A₃, calystegine B₂, and calystegine B₃, and N-alkyl derivatives thereof.

6. The method of claim 5 wherein said compound is 1-deoxygalactonojirimycin or 3,4-diepi- α -homonojirimycin.

7. The method of claim 6 wherein said compound is 1-deoxygalactonojirimycin.

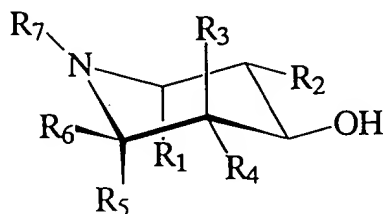
8. A method of enhancing the activity of lysosomal α -galactosidase A in mammalian cells comprising administering an effective amount of a compound of the formula



wherein

- R_1 represents H, $-\text{CH}_2-$ or CH_2OH ;
- R_2 represents H, OH or $-O$ -galactose;
- R_3 and R_4 independently represent H, or OH;
- R_5 represents H, or $-\text{CH}_2-$;
- R_6 represents CH_2OH , or OH; and
- R_7 represents H or an alkyl group containing 1-3 carbon atoms, provided that when either R_1 or R_5 is $-\text{CH}_2-$, they are identical and are linked to form a second ring structure.

9. A method of treating Fabry disease comprising administering an effective amount of a compound of the formula



wherein R₁ represents H, -CH₂- or CH₂OH;

R₂ represents H, OH or -O-galactose;

R₃ and R₄ independently represent H, or OH;

R₅ represents H, or -CH₂-;

R₆ represents CH₂OH, or OH; and

R₇ represents H or an alkyl group containing 1-3 carbon atoms, provided that when either R₁ or R₅ is -CH₂-, they are identical and are linked to form a second ring structure.